

Psychosocial Management of the Patient With Duchenne Muscular Dystrophy

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abstract

In this article, we outline a comprehensive plan for the psychosocial management of patients with Duchenne muscular dystrophy (DMD) across the life span. In 2010, the Centers for Disease Control and Prevention sponsored the development of multidisciplinary management guidance for DMD, and in 2018, that guidance was updated. In the intervening years, a new emphasis was placed on studying and addressing the psychosocial issues that affect patients with DMD, driven in part by improved patient survival. Once viewed as ancillary to managing the significant medical needs of patients with DMD, it is now standard practice to integrate psychosocial management into the multidisciplinary management of the disease. It is also increasingly recognized that neurodevelopmental disorders in DMD occur at a higher rate than what was previously understood and that these disorders warrant early and intensive intervention. In this article, we expand on the content found in the 2018 DMD Care Considerations.

Significant progress has been made in managing the medical needs of patients who have Duchenne muscular dystrophy (DMD). As a result, many patients live well into adulthood and have the potential to lead independent, productive lives. However, the challenge of coping with a chronic and progressive illness from childhood into young adulthood is formidable. Close monitoring and appropriate interventions focused on improving emotional and social functioning should be

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considered integral elements of multidisciplinary DMD management. A developmental perspective is required to support the gradual transition of medical management and decision-making from parents to patients as well as a systems perspective emphasizing family and community involvement. This approach is especially critical because patients with DMD may be at an increased risk of cognitive and social developmental delays.¹⁻³ This is an emerging area that is ripe for future research to develop interventions specific to DMD that can be integrated across medical settings. In this article, we discuss what is currently known about the psychosocial health of patients with DMD, identifying areas that warrant screening and monitoring and discussing the state of evidence-based interventions to address areas of concern.

A major impetus for this work is that certain genetic variants associated with DMD are now known to result in atypical dystrophin expression in the brain.²⁻⁴ Compared with healthy children, patients with DMD have smaller total brain volume, reflecting lower cortical (gray matter) volume and atypical white matter connectivity.⁵ These brain characteristics may contribute to the relatively high rate of neuropsychiatric disorders in patients with DMD. In recent studies of patients with DMD, approximately 32% of patients had attention-deficit/hyperactivity disorder (ADHD), 26% had learning difficulties, 17% to 27% had an intellectual disability, 27% had anxiety, and 15% had autism spectrum disorder (ASD).¹⁻³ Specific learning disabilities that affect language-based academic skills may be more common.⁶ Cognitive strengths and limitations may be evident on neuropsychological testing,⁷⁻⁹ which should be considered for patients with DMD

who are suspected of having a superimposed neurodevelopmental disorder. In such cases, early intervention is critical in minimizing the compounding impact of any delays over time.

In addition, it has long been recognized that patients and their family members are at an increased risk of depression and anxiety, especially as the disease progresses.¹⁰ These factors may interfere with optimal medical management and contribute to a poorer quality of life. Routine screening for these concerns may reduce stigma and delays in the provision of appropriate mental health treatment.

CORE RECOMMENDATIONS

Given the significant needs of patients with DMD, most patients will receive care in neuromuscular clinics within major tertiary-care hospitals that allow for coordinated care across specialties. Many patients also will receive some degree of care within their local communities. All providers should be mindful of the psychosocial functioning of their patients with DMD and the impact of the disease on their families. Providers can play an active role in developing and implementing psychosocial supports across their patients' life spans. Helping patients to think about the future and empowering them to actively participate in their own care and daily activities constitute components of this psychosocial support.

Pediatricians and primary care physicians who care for patients with DMD may be the first clinicians to identify psychosocial concerns. (For additional details, see the primary care and emergency department specialty article that is part of this supplement.¹¹) Within the hospital-based clinic, a care coordinator should be available, acting as a link to community- and

hospital-based care and serving as a primary point of contact for the management of emotional and social stressors. Psychosocial health should be assessed regularly in the neuromuscular clinic through the use of screening tools. If concerns are identified, then more intensive psychiatric services may be warranted. The care providers who should be involved in this process and their individual roles are outlined in Table 1. Core psychosocial care considerations organized by stage of disease are shown in Fig 1.¹²

Care Coordination

A care coordinator should be embedded in the neuromuscular clinic to serve as the primary point of contact for patients and families. The coordinator should have a medical background (eg, as a nurse or nurse practitioner), be familiar with neuromuscular disorders, and have close connections with relevant specialists. The care coordinator may be the first provider to identify and address psychosocial needs of a patient and family, providing the family with routine clinical information and facilitating referrals to mental health providers and developmental specialists when needed.

Psychological Management

The DMD care team should include a mental health clinician (psychologist, psychiatrist, social worker, or psychiatric nurse) who has specialized training and experience in working with patients and families affected by chronic medical or neurodevelopmental conditions. At each visit to the neuromuscular clinic, mental health and quality of life should be screened by using standardized questionnaires. An appropriate tool for pediatric patients is the Strengths and Difficulties Questionnaire,¹³ which is available in multiple languages. For adult patients, the

TABLE 1 Components of Clinic-Based Psychosocial Care of Patients With Duchenne Muscular Dystrophy

Care coordination	
The care coordinator is a point of contact for patients with Duchenne muscular dystrophy (DMD) and families; they should be health professionals with sufficient training or experience in the clinical care of patients with DMD	
The role of the care coordinator is to provide information, coordinate (and possibly schedule) appointments, and facilitate communication with clinicians across disciplines	
Routine mental health screening	
At each neuromuscular clinic visit, mental health and quality of life should be screened	
Screening can be informal and does not require comprehensive assessment	
An appropriate tool for paediatric patients is the Strengths and Difficulties Questionnaire; for adult patients, the Patient Health Questionnaire 9-item depression scale (PHQ-9) and the Generalized Anxiety Disorder 7-item scale (GAD-7) are appropriate; for parents of patients aged 5–17 years, the Personal Adjustment and Role Skills Scale (PARSIII) is suitable (scale and scoring programme is available on the Parent Project Muscular Dystrophy website)	
Screening can be conducted by a social worker or mental health professional or by other clinic staff with sufficient training or experience in this area (eg, a nurse or attending physician)	
If screening is positive, a referral should be made to a psychologist and psychiatrist for further assessment or treatment	
Every clinic should have a plan to assess and address suicidal ideation or other acute safety concerns	
Caregiver emotional adjustment should be monitored and intervention or support offered as needed	
Siblings of a person with DMD should be provided with opportunities to connect with other siblings of patients with DMD and with access to mental health services as needed	
Psychological care	
All individuals with DMD should be expected to live rich, fulfilling lives, and those without additional neurodevelopmental or psychological disorders may achieve a high level of independence in managing their disease; however, all patients and their families might need psychosocial support	
The neuromuscular care team should include a mental health professional (ie, psychologist or psychiatrist) with training and experience in assessing and treating psychiatric conditions in the context of chronic medical or neurodevelopmental conditions	
When mental health concerns are identified, the mental health professional should provide further evaluation of individuals with DMD and their family members, and provide cognitive or behavioral interventions to treat psychiatric conditions	
Standard, evidence-based practices should be used for those who need more formal mental health treatment	
Neuropsychological evaluations should be done when cognitive delays, difficulties with emotional and behavioral regulation, or concerns about social skills exist; re-evaluations should be done every 2–3 years to monitor developmental progress and response to interventions	
Neuropsychological evaluations should be considered within the first year of diagnosis to establish a baseline, or when transitioning to adulthood if government-based assistance might be necessary to promote functional independence	
Pharmacological interventions	
The neuromuscular team should include a psychiatrist or other physician with training and experience in providing medication to treat behavioral or emotional disorders in the context of chronic medical or neurodevelopmental conditions	
Standard prescribing practices should be followed	
Selective serotonin-reuptake inhibitors should be prescribed for depression, anxiety, and obsessive–compulsive disorder	
α-Adrenoceptor agonists (first choice) or atypical antipsychotics (second choice) should be prescribed for aggression and anger or emotional dysregulation	
Stimulants or α-adrenoceptor agonists should be prescribed for attention-deficit hyperactivity disorder	

GAD-7, Generalized Anxiety Disorder 7-item scale; PARSIII, Personal Adjustment and Role Skills Scale; PHQ-9, Patient Health Questionnaire 9-item scale. Reprinted with permission from Birnkrant DJ, Bushby K, Bann CM, et al; DMD Care Considerations Working Group. Diagnosis and management of Duchenne muscular dystrophy, part 3: primary care, emergency management, psychosocial care, and transitions of care across the lifespan. *Lancet Neurol*. 2018;17(5).

Patient Health Questionnaire is recommended (Patient Health Questionnaire-9 for depression¹⁴; Generalized Anxiety Disorder-7

for anxiety¹⁵). Finally, for parents of patients ages 5 to 17 years, the Personal Adjustment and Role Skills Scale is recommended (the scale and

scoring program is available on the Parent Project Muscular Dystrophy Web site: www.parentprojectmd.org/ForClinicians).¹⁶ Screening scores should be entered into the medical record to allow tracking of symptoms over time. If the screening scales are positive, then further evaluation by the mental health clinician is warranted. The mental health clinician should work with each patient and family to develop a safety plan, and all DMD clinics should have a protocol in place to address patient suicidal ideation.

Because screening measures have limited sensitivity, the mental health clinician should conduct an annual formal mental health assessment on each patient in the DMD clinic. This longitudinal assessment gives the mental health clinician the opportunity to build a professional relationship with the patient and the family, which is an asset if the patient's mental health care needs intensify over time. When formal treatment of psychological disorders is necessary, then standard, evidence-based practices should be used, ideally involving practices that have been validated for populations with chronic medical conditions. For example, if patients present with significant symptoms of anxiety, then cognitive behavioral therapy (CBT) may be appropriate, but therapists may also need to integrate behavioral medicine approaches to understand fully how psychological symptoms manifest in the context of a chronic medical condition.

Clinicians should consider psychopharmacologic interventions when treating moderate to severe psychiatric symptoms and when treating milder symptoms that are not amenable to nonpharmacologic interventions. Standard prescribing practices and guidelines apply and are subject to the patient's age and medical condition. For example, special attention must be paid to medication interactions, the patient's

	Ambulatory stage or childhood	Early non-ambulatory stage, adolescence, or young adulthood	Late non-ambulatory stage or adulthood
Assessments	<div><div><div>• Consider baseline evaluation during first year of diagnosis</div><div>• Provide developmental (<4 years old) or neuropsychological evaluation (>5 years old) when social or emotional concerns or cognitive delays exist</div></div><div>Provide evaluation by speech-language pathologist for children with suspected delays in speech or language development</div><div>Provide evaluation by social worker at diagnosis and then as needed</div></div> <div></div> <div></div>	<div><div><div>• Provide neuropsychological evaluation to identify cognitive or learning issues when concerns exist about school performance</div><div>• Provide neuropsychological evaluation when transitioning to adulthood to assess whether government-based assistance might be needed</div></div><div></div><div>Provide evaluation by social worker of the needs of the patient and family</div></div> <div></div> <div></div>	<div><div>Provide neuropsychological evaluation when concerns exist about change in functioning or ability to manage daily affairs</div><div></div><div>Provide evaluation by speech-language pathologist for patients with loss or impairment of functional communication ability, chewing difficulties, or dysphagia</div></div> <div></div> <div></div>
Interventions	<div>Refer for psychotherapy or psychopharmacology, or both, when mental health concerns are identified for the patient or family</div> <div><div>Implement formal accommodations at school for health, safety, and accessibility; plan for health-related absences</div><div>Set goals for future education and vocation</div></div> <div><div>Provide parents with resources to educate teachers, school psychologists, and other school personnel about DMD</div><div>Provide parents and patients with resources to educate peers about DMD</div><div>Refer to psychologist for social skills training as needed</div></div> <div>Encourage patients and families to stay active and engaged</div> <div>Promote patient self-advocacy and independence</div> <div></div> <div></div> <div></div>	<div></div> <div></div> <div></div> <div></div> <div></div> <div></div> <div></div> <div></div> <div></div> <div></div> <div></div> <div></div> <div></div> <div></div> <div></div> <div></div> <div></div> 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FIGURE 1

Care considerations for the psychosocial care of individuals with DMD. (Reprinted with permission from Birnkrant DJ, Bushby K, Bann CM, et al; DMD Care Considerations Working Group. Diagnosis and management of Duchenne muscular dystrophy, part 3: primary care, emergency management, psychosocial care, and transitions of care across the lifespan. *Lancet Neurol*. 2018;17[5].)

cardiac status, and potential side effects of medication, such as weight gain. Because psychiatric presentation and relevant medical and psychosocial issues change over

time, close monitoring and regular follow-ups are highly recommended. Cardiac disease is of special concern because its progression over time may affect the decision to use various

behavioral medications, including stimulants, atypical antipsychotics, and some antidepressants. When such medications are being considered, close collaboration

is needed between specialists in psychiatry, cardiology, and neurology to help patients and their families deal with the complex risk/benefit assessments that are involved.

Neuropsychological Evaluations

A comprehensive psychological assessment should be conducted when concerns that warrant possible treatment are identified. Because of the prevalence of learning issues and the frequency of comorbidities that may complicate diagnosis and treatment (eg, ADHD in combination with both anxiety and learning disabilities), the assessment should include neuropsychological testing. Testing should be conducted by a neuropsychologist, who is a clinical psychologist with specialized training and experience in assessing neurologic and psychiatric disorders. Neuropsychological assessment includes standardized testing tailored to the patient's developmental stage, assessing the patient's current cognitive functions, academic skills (or school readiness for younger patients), social functioning, emotional adjustment, and behavioral regulation. With family input, the results of the assessment can be used to create targeted recommendations, including an intervention plan that can be implemented at home and in school. Comprehensive reevaluations are often warranted every 2 to 3 years to monitor the child's developmental progress and response to interventions. Circumstances that require more frequent reevaluations include an acute change in functioning, new psychological concerns, or a major transition at home or in school.

SPECIFIC TARGETS FOR INTERVENTION BY DEVELOPMENTAL AND DISEASE STAGE

In the following sections, we present guidance for psychosocial management organized by stage

of disease: for children in the ambulatory stage (Table 2), adolescents in the early nonambulatory stage (Table 3), and adults in the late nonambulatory stage of DMD (Table 4).

Childhood: Ambulatory Stage

At the time of initial diagnosis, patients and families should have regular contact with the care coordinator, who can facilitate visits with specialists and provide appropriate information about local and online resources and connections to national advocacy groups.¹⁷ The mental health clinician should also perform a formal assessment of the patient, caregivers, and other family members. Within the first 12 months after diagnosis, a comprehensive neuropsychological evaluation is recommended to establish a baseline to monitor developmental progress and determine if there is evidence of comorbid neurodevelopmental conditions or uneven development of cognitive skills that may require remediation.

Psychological Management

Caregivers generally report a high level of life satisfaction,^{18,19} but they also report stress and emotional and physical burden.²⁰ Providing support to caregivers at the time of diagnosis is critical in promoting their successful adjustment. Physicians and the care coordinator should be in regular contact with caregivers at this stage and project a message of hope as well as caring, empathy, and teamwork.^{17,21} Caregiver interventions focused on enhancing the mother's resilience and social support as well as enhancing positive perceptions of the impact of disease have been found to be the most beneficial.^{21–23} Genetic counseling also may play an important role but should be done with consideration of the psychological impact of a carrier diagnosis on the mothers and grandmothers of children with DMD, which may include profound,

long-lasting guilt and a strong sense of responsibility for the genetic variants' occurrence in the family.^{24,25} Testing for carrier status after a period that allows for the family to adjust to the initial diagnosis may help mitigate feelings of self-blame.

Around the time of initial diagnosis, attention should be paid to helping parents and family members adjust to a diagnosis of DMD and develop longer-term coping strategies. However, the child's adjustment should also be considered, including the child's ability to understand and cope with the emotional turmoil that inevitably occurs in the family at the time of diagnosis. Families should be encouraged to discuss DMD with the child as early as possible but in a way that matches the child's developmental maturity. Starting the conversation during childhood helps to normalize discussion of DMD, sets an expectation that the child will participate in medical decision-making, and potentially facilitates transition to functional independence as the child grows older. The mental health clinician should offer ongoing advice and resources to parents to support this process.

In younger children, the strength of social networks and opportunities for friendships have been shown to be associated with resilience.²⁶ However, social functioning may be an area of primary concern because children with DMD may struggle to interact with peers and develop friendships. The risk of social difficulties may be the result of limitations in physical activities and play, although immature behavior and interests and excessive shyness or social anxiety may be present at higher rates.^{27,28} Social-skills training is critical for these patients, as are interventions that target behavioral regulation and anxiety. Anxiety may not always manifest in the traditional sense (eg, excessive worry); it can be characterized by rigid expectations and overly concrete reasoning, which cause the

TABLE 2 DMD Assessments and Interventions for Childhood (Ambulatory Stage)

Assessment and Intervention	Considerations
Psychosocial assessments	
Neuropsychological assessment	<ul style="list-style-type: none"> • A neuropsychologist is a doctoral-level clinical psychologist with expertise in brain functioning and development within the context of medical conditions. • Comprehensive developmental (children <4 y old) or neuropsychological (children >5 y old) assessment to (1) characterize cognitive, emotional, social, and academic skills; (2) develop a plan to address these areas of functioning; and (3) diagnose comorbid neurodevelopmental conditions (eg, ASD, learning disabilities, and ADHD). • Evaluation should be near the time of diagnosis and/or before entering formal schooling and then every 2–3 y to monitor developmental progress or when there are concerns about changes in developmental trajectory.
Speech and language	<ul style="list-style-type: none"> • Evaluation of expressive and receptive language skills by a certified speech and language pathologist is necessary for younger children who present with suspected delays in speech and/or language development (as identified by the caregiver or because of professional's concerns). The speech and language therapist may also develop and execute a treatment plan.
Social worker	<ul style="list-style-type: none"> • A social services professional is defined as a clinical social worker or other professional who (1) is sufficiently trained and qualified to assess and address emotional adjustment and coping, (2) has knowledge and/or access to financial resources and assistance programs, and (3) has access to social support networks. This professional should have an understanding and/or experience with DMD. • Evaluation should be near the time of diagnosis and on an ongoing basis that is determined by the needs of the patient and family. • Disease self-management: developmentally appropriate awareness of diagnosis, medications, and medical management.
Psychotherapy	<ul style="list-style-type: none"> • Parental management training: recommended for externalizing behaviors (eg, noncompliance, disruptive behavior, and parent–child conflict). • Individual therapy: CBT is recommended for internalizing conditions (eg, low self-esteem and depression, anxiety disorders and OCD, and adjustment and coping difficulties). • Group therapy: recommended for social skills weaknesses • Family therapy: recommended for adjustment and coping difficulties and parent-child conflict.
Educational interventions	<p>At a minimum, formal accommodations should be specified at school to allow for the following:</p> <ul style="list-style-type: none"> • modifications of activities that might be harmful to muscles (eg, adaptive physical education); • compensation for reduced energy or fatigue (eg, walking long distances to and from lunch); • addressing safety (eg, playground activities); • addressing accessibility (eg, assistive technology and going up stairs); • planning in advance for medically related absences (eg, doctor's appointments and home tutoring in the event of an extended absence); • all accommodations should be as inclusive as possible. <p>Special education services should be provided to children and adolescents with additional learning, behavioral, and psychosocial concerns.</p> <p>Increase DMD awareness and knowledge among school personnel.</p>
Social development	<ul style="list-style-type: none"> • Peer education about DMD • Social skills training (as needed) • Encourage patients and families to stay active and engaged (modified and/or adapted sports, summer camps, youth groups or programs, patient advocacy groups, and use of service dogs). • Promote patient self-advocacy and independence.

child to have difficulty in adjusting to unexpected circumstances or changes in routines. Behavioral rigidity can trigger agitation and emotional reactivity, including oppositional behavior, arguing, and emotional outbursts. These behaviors may reflect neurologically based weaknesses in certain problem-solving skills rather than purposeful attempts to manipulate others. They also may be the result of inconsistent or overly permissive parenting. In these cases, behavioral therapy may be warranted, including training that

is focused on equipping parents with strategies that they can implement across settings. Such training increases compliance, de-escalates emotional outbursts or conflict, and promotes better problem solving. Psychological care is particularly important for children who have comorbid neurodevelopmental or neuropsychiatric conditions (eg, ASD, ADHD, anxiety, or obsessive-compulsive disorder [OCD]). For these children, standard treatment practices, including therapeutic techniques and medication, should

be employed, although modifications may be necessary because of the impact of DMD on psychosocial functioning. For example, for the child who has both DMD and ASD, applied behavior analysis therapy may be appropriate, but goals may relate to compliance with medications and other therapies. Similarly, when considering medication to treat ADHD, the patient's overall functional and medical status should be considered. Early identification and treatment is important for optimizing the

TABLE 3 DMD Assessments and Interventions for Adolescence (Early Nonambulatory Stage)

Assessment and Intervention	Considerations
Psychosocial assessments	
Neuropsychological assessment ^a	<ul style="list-style-type: none"> • Reevaluation to identify issues in cognition and learning, with changes in school performance or parent and/or teacher concerns.
Social worker ^a	<ul style="list-style-type: none"> • Evaluation of the needs of the family and child; this should include changes brought about by physical changes (equipment needs), social needs (social isolation or community involvement), emotional needs (depression, anxiety, and loneliness), and developmentally appropriate issues (independence, decision-making, relationships, sexuality, and risk-taking behaviors).
Psychotherapy	<ul style="list-style-type: none"> • Parental management training: recommended for externalizing behaviors (eg, noncompliance, disruptive behavior, and parent-child conflict); support for adolescent independence, decision-making, setting limits, avoiding social isolation, school and/or community engagement, future goal setting. • Individual therapy: CBT is recommended for internalizing conditions (eg, low self-esteem and depression, anxiety disorders and OCD, and adjustment and coping difficulties). • Group therapy: recommended for social skills weaknesses. • Family therapy: recommended for adjustment and coping difficulties and parent-child conflict. • Disease self-management: developmentally appropriate awareness of diagnosis, medications, medical care and management, and assistive devices; begin to take responsibility for care and management.
Educational interventions	<p>At a minimum, formal accommodations should be specified at school to allow for the following:</p> <ul style="list-style-type: none"> • Modifications of activities that might be harmful to muscles (eg, strenuous physical education), • Compensation for reduced energy or fatigue (eg, walking long distances to and from lunch), • Addressing safety (eg, playground activities), • Addressing accessibility (eg, assistive technology and going up stairs), • Planning in advance for medically related absences (eg, doctor's appointments and home tutoring in the event of an extended absence), and • Beginning to set goals for future education and/or vocation; identifying plans to meet these goals; all accommodations should be as inclusive as possible. <p>Special education services should be provided to children and adolescents with additional learning, behavioral, and psychosocial concerns.</p> <p>Increase DMD awareness and knowledge among school personnel.</p>
Social development	<ul style="list-style-type: none"> • Peer education about DMD. • Social skills training (as needed). • Encourage patients and families to stay active and engaged (modified and/or adapted sports, summer camps, youth groups or programs, patient advocacy groups, and use of service dogs). • Promote patient self-advocacy and independence. • Social interaction interventions: should be provided with sex education that includes information about sexuality and disabilities, inheritability of genetic conditions, etc. • Care and/or support interventions: transition planning: encouraging self-advocacy in medical care, facilitating transfer to a new medical care team, and developing educational and vocational opportunities.

^a See Table 2 for definition.

patient's quality of life and family functioning.

Neuropsychological Evaluations

Awareness is growing that neurodevelopmental or cognitive difficulties in young patients with DMD correlate with later cognitive functioning, making early identification of these difficulties important.²⁹ Even in preschool-aged patients, neuropsychological evaluations may identify cognitive difficulties^{30,31} as well as difficulties with social and behavioral regulation that may manifest as reduced adaptive functioning.³¹ On the basis of neuropsychological testing results, specialized evaluations and

interventions may be warranted. Evaluations may be repeated over the course of childhood and adolescence to monitor developmental functioning and how the child responds to interventions.

Special Education Needs

Children with DMD should have an individualized academic plan. At a minimum, accommodations are needed to promote integration with peers who are not affected while recognizing physical limitations. Common accommodations protect the child's muscles (eg, avoidance of strenuous physical activities), reduce fatigue (eg, avoidance of long walks between classes), and increase

safety and accessibility (eg, use of assistive technology). Academic accommodations are necessary to address medically related absences (eg, tutoring or adjustments to workload). Children and adolescents who have additional neurodevelopmental concerns (eg, ADHD, ASD, or dyslexia) may need additional special education services to ensure consistent access to the academic curriculum.

In the United States, access to special education accommodations and services is mandated by federal legislation under the Individuals with Disabilities Education Act. For school-aged children, academic accommodations should be formalized as part of a 504 plan

TABLE 4 DMD Assessments and Interventions for Adulthood (Late Nonambulatory Stage)

Assessment and Intervention	Considerations
Neuropsychological assessment	<ul style="list-style-type: none"> Evaluations may be spaced further apart in adulthood but should be performed when there are concerns about a change in functioning (cognitive, emotional, and behavioral) or when specific concerns emerge regarding the ability to manage daily affairs.
Speech and language consultation	<ul style="list-style-type: none"> Consider for older patients who present with loss or impairment of functional communication ability Refer patients having issues with chewing or dysphagia.
Social work	<ul style="list-style-type: none"> Encourage community and family engagement, peer activities, independence, patient advocacy groups, and use of service dogs. Promote patient self-advocacy and independence. Facilitate mentoring or support programs from which young men with DMD can obtain information, advice, and support from other men with DMD (eg, vocational development, relationships, and parenting). Disease self-management: awareness of diagnosis, medications, medical care and management, and assistive devices; full responsibility for medical care and management.
Psychotherapy	<ul style="list-style-type: none"> Individual therapy: CBT is recommended for internalizing conditions (eg, low self-esteem and depression, anxiety disorders and OCD, and adjustment and coping difficulties). Group therapy: recommended for social skills weaknesses. Family therapy: recommended for adjustment and coping difficulties and parent-child conflict
Educational interventions	<ul style="list-style-type: none"> Assistance with continuing education, vocational training, employment; extended transitional education with an IEP (in the United States) until 22 y of age. Assistance with adjustments in care routines, care assistance, and to accommodate job requirements.
Care and/or support interventions	<ul style="list-style-type: none"> Home health care services should be used if a patient's health is at risk because sufficient care cannot be provided in their current setting or circumstances; it might also be appropriate in other situations when current care providers cannot sufficiently meet the patient's care needs. Notify patients and families about the availability of palliative care: appropriate for managing pain, clarifying treatment goals and difficult medical decisions, facilitating communication between patients and families and medical teams, providing emotional and/or spiritual support, and addressing grief, loss, and bereavement. Assist with arranging respite care for caregivers, especially when patients are ventilated. Make hospice care available for end-stage patients.

or in an Individualized Education Program (IEP). Details regarding these different plans are available from local and state departments of education (see, for example, information from New Hampshire's Department of Education). Educational evaluations also are available through the local public school system by parent request and may be used to identify whether a child is struggling in a particular area. Neuropsychological evaluations should be performed to understand the underlying contributing factors in the context of a child's medical and developmental history and to develop a more comprehensive treatment plan that includes, but is not limited to, academic functioning.

Adolescence: Early Nonambulatory Stage

During adolescence, children with DMD should begin to acquire skills that promote independence at a rate that matches the child's level of cognitive, emotional, and social

functioning. During early adolescence, care providers should start to prepare patients with DMD for autonomous medical decision-making. This will allow time for children with DMD to acquire experience in managing their own care so that when the transition to adulthood arrives, it is as seamless as possible.

Psychological Management

Many patients with DMD will face typical challenges commonly associated with adolescence, but the nature of these challenges will be influenced by the disease. Adolescents with DMD may be at an increased risk for depression and anxiety,³² especially if there is a decline in quality of life. The physical demands of the disease increase over time, resulting in chronic pain and fatigue in adolescence and adulthood.^{33–35} The loss of ambulation reduces independence and may cause the patient to become self-conscious and isolated from peers. Young adults may have problems related to social

development at a stage in life when developing personal relationships (romantic and otherwise) becomes more of a priority.

The family is also at an increased risk of psychological stress during this transition phase. Parents are more likely to develop depression when their child with DMD reaches adolescence,³⁶ especially when the child loses ambulation.^{10,20} Parents may miss symptoms of depression in their child at a time when the child's quality of life is declining.³⁷ Siblings are also at risk for psychological distress, especially when the patient becomes more dependent on the family for activities of daily living.³⁸ Clinical management at this stage should be used to address the impact of the disease on parents and siblings, focusing on resilience and social supports.²² Greater involvement of siblings may mitigate family distress,³⁹ but efforts to involve the family also should respect the patient's age-appropriate desire for greater autonomy.

Special Education Needs

The continuation of academic supports for postsecondary education may be warranted. In late adolescence, discussion and planning related to the transition to adulthood should occur, including developing skills to support the greatest degree of functional independence and a vocation. For patients who have IEPs, services may be provided until age 22 years old.

Adulthood: Late Nonambulatory Stage

Psychological Management

Many people living with DMD are psychologically well adjusted,^{26,37} pursuing secondary education and careers, establishing significant relationships, and having children. Discussions related to sexuality should be raised in adolescence and continued into adulthood. Similarly, patients and their partners may wish to consider genetic counseling to obtain information and guidance about family planning. Chronic pain and fatigue should be continuously managed because these increase the risk for anxiety and depression,⁴⁰ which may become more frequent in adulthood.³² Invasive ventilation, if employed, can significantly reduce health-related quality of life⁴¹ and is associated with greater caregiver burden.^{18,42} Subjective caregiver burden is alleviated by support and respite care as well as intervention programs that alleviate anxiety and foster coping skills.

Neuropsychological Evaluations

Evaluations may be spaced farther apart in adulthood but should be performed when concerns about a change in functioning (cognitive, emotional, or behavioral) emerge or if the patient has specific concerns about the ability to manage daily affairs. Evaluations are also sometimes required to determine a patient's eligibility for state-funded services.

Vocational Supports

As patients transition to the workplace, they also may need

support in adjusting their care routines to job requirements. Patients who have cognitive or neurodevelopmental disorders may need formal vocational training, which, in the United States, may be available from state-funded programs. Such training should ideally begin in secondary education. In the United States, all employment locations are required to provide accessibility on the basis of the Americans with Disabilities Act of 1990.

Palliative Care

As the disease progresses, palliative care should be integrated into clinic visits. Recent studies have revealed that few patients or families have an awareness of palliative care services.⁴³ In addition to pain management, palliative care teams may be able to provide emotional and spiritual support, assist families in clarifying treatment goals and making difficult medical decisions, facilitate communication between families and medical teams, and address issues related to grief, loss, and bereavement. The neuromuscular clinic's care coordinator should help patients and their families access palliative services.

EMERGING AREAS AND FUTURE DIRECTIONS

Patients with DMD are now expected to survive into adulthood, making psychosocial care more important because it has a direct impact on the quality of life of patients and their families. Studies are needed to elucidate the rates and etiology of comorbid neurodevelopmental conditions, including ASD and learning disabilities. Anxiety appears to be a prevalent problem that has not been examined in a comprehensive manner. Widespread adoption of mental health screenings and scheduled neuropsychological assessments will help to establish the efficacy of interventions in improving

quality of life and medical functioning at different developmental stages.

On a basic research level, studies have only just begun to recognize that genetic mutations may play a role in the expression of dystrophin in the brain and to describe how this influences the development of the central nervous system. The degree to which dystrophin expression selectively affects certain brain systems and how this relates to cognition and behavior is still unclear. The psychosocial implications of carrier status are also not fully understood. Emerging research has been focused on the implications of female carrier status on cardiac and musculoskeletal function,⁴⁴ but researchers also recognize that carrier status may entail greater risk for cognitive and psychiatric disorders, an area that remains largely unexplored.^{45,46}

Studies have consistently revealed that DMD places a significant burden on the family.²⁰ These burdens can be financial, physical, and emotional and can affect the patient's caregivers, parents, and siblings. Advances in psychosocial management will reduce these burdens and improve quality of life. The benefits of effective psychosocial evaluation and management of patients with DMD and their families cannot be overstated, and additional research is greatly needed in this important area of management.

ABBREVIATIONS

ADHD:	attention-deficit/hyperactivity disorder
ASD:	autism spectrum disorder
CBT:	cognitive behavioral therapy
DMD:	Duchenne muscular dystrophy
IEP:	Individualized Education Program
OCD:	obsessive-compulsive disorder

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